BENIGN NEUROGENIC TUMORS OF THE CHEST WALL*

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WITH THE ADVENT OF mass roentgenography of the chest in recent years, and the increased proficiency of thoracic surgery, more intrathoracic tumors have been subjected to surgical removal than in former years. Approximately 200 cases[1, 2, 12, 19, 20] of benign intrathoracic neurogenic tumors have been recorded in the literature. A few benign neurogenic tumors have been found in the anterior mediastinum,[19] and also in the lung,[27] but classically these tumors arise from the neural elements in the vicinity of the paravertebral gutter. Also in the latter location are those tumors which arise from the sympathetic nervous system (*i.e.*, ganglioneuroma). Only on rare occasions have benign tumors of the intercostal nerves been found arising in other parts of the chest wall. A review of the literature reveals reports of only 18 benign nenurogenic chest wall tumors which were removed surgically (Table I) exclusive of those involving the mediastinum. We have encountered four similar tumors and are reporting them with a resumè of the other 18 cases.

Incidence. Well over 500 chest wall tumors have been reported[4, 7, 13, 14, 18, 25, 28] in the surgical literature. The majority of these tumors arise from the osseous elements of the thorax and about half of them are sarcomatous.[16] In the other tumors of the chest wall, neurogenic tumors comprise about one-third of the total number of cases, with neurogenic sarcomas outnumbering the benign neurogenic tumors by about ten to one.

Pathology. Benign tumors arising from peripheral nerves can originate from the connective tissue sheath of a nerve or from the nerve fibers themselves. In the latter group there are no true neoplasms, the tumor usually arising locally after injury, as exemplified by the "amputation neuroma." The classification and histogenesis of the benign tumors arising from the mesodermal elements of a nerve has been the subject of considerable disagreement. Some authors prefer to group all these tumors in the one category of "neurofibroma," while others have classified them as "perineural fibroblastoma,"[12] reserving the term neurofibroma for those tumors which are associated with von Recklinghausen's disease—neurofibromatosis. Basically, there are two distinct histologic forms of benign neurogenic tumors,[9, 10, 26] with the neurofibroma being the commonest type (Fig. 1). Grossly, this tumor is not encapsulated, grey in color and fairly firm in consistency. Histopathologically, it consists of elongated Schwann cells interspersed between haphazardly arranged connective tissue cells (Fig. 1). In general, these tumors tend to resemble a cellular fibroma.

It is characteristically found in von Recklinghausen's disease, where one also encounters cutaneous pigmentation and other small cutaneous neurofibromas. Not infrequently, a solitary neurofibroma of the chest wall is found unassociated with generalized

* Submitted for publication December, 1952.

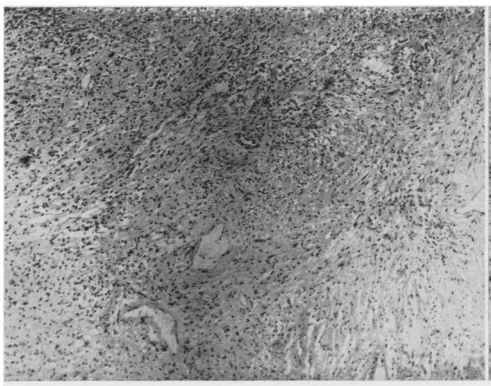

FIG. 1

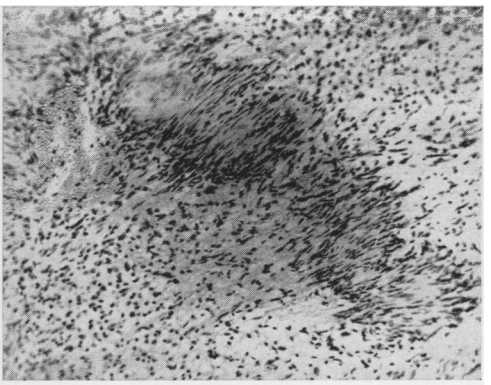

FIG. 2

FIG. 1. Photomicrograph demonstrating interlacing network of cells and fibers in disorderly arrangement, characteristic of a neurofibroma.

FIG. 2. Photomicrograph of a neurilemoma demonstrating palisading of elongated Schwann cells.

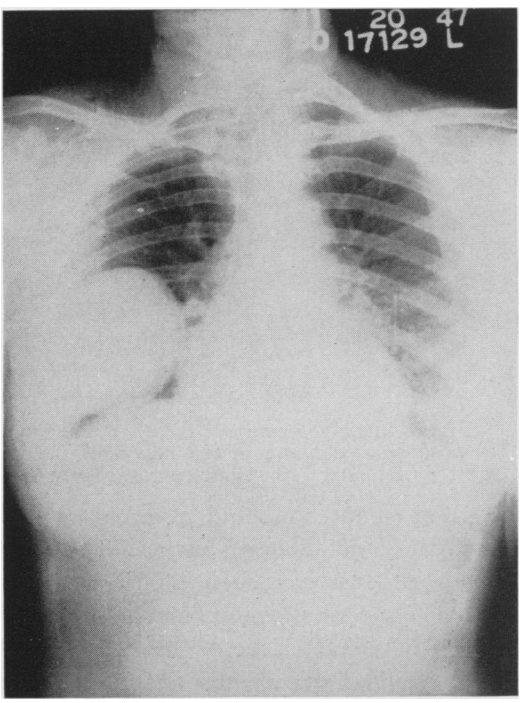

FIG. 3. There is a large circumscribed mass in the lower lateral part of the right hemithorax. The tumor extends from above the seventh rib to below the tenth rib posteriorly. Same case as Figure 4.

neurofibromatosis. Von Recklinghausen's disease was present in only two of the 22 cases reported herein. The first chest wall neurofibroma was reported by Mouchet[21] in 1900. In this case, a large neurofibroma of an intercostal nerve, multiple small nodules in other intercostal nerves and a glioma of the fourth ventricle were found at necropsy.

The second histologic type of benign nerve tumor is the neurilemoma (Fig. 2). This tumor is occasionally referred to as a Schwannoma, neurinoma and perinueral fibroblastoma. It is usually well encapsulated, yellowish in color and frequently contains areas of necrosis and degeneration, especially when the tumor has attained large size. These tumors are usually solitary growths and are not associated with von Recklinghausen's disease. Histologically they are solid in appearance and present an interlacing syncytium of Schwann cells with prominent palisading of the cell nuclei (Fig. 2). Only five of the 22 chest wall tumors were classified as neurilemomas, although with more uniform histopathologic classification, the number might be increased. The neurilemoma is probably always benign and does not recur after complete surgical removal. By contrast, recurrence is not uncommon after surgical excision of the neurofibroma and malignant transformation in the latter type of tumor is quite common. It has been estimated that 18 per cent of all neurofibromas show malignant changes. Undoubtedly, a number of neurogenic sar-

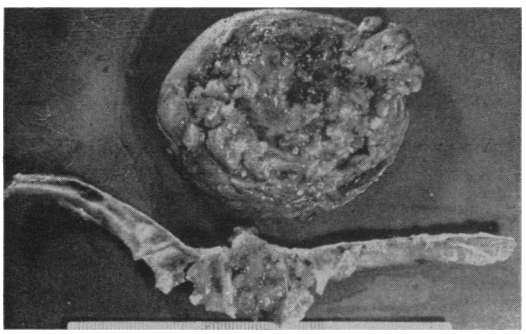

FIG. 4. Tumor and rib removed from patient whose roentgen ray is shown in Figure 3.

comas of the chest wall originated in benign neurofibromas at that site.

Clinical Manifestations. In this small series of 22 cases of benign neurogenic tumors of the chest wall there is no significant sex predisposition. The ages of the patients ranged from 17 to 67 years. There was no significant predilection for site of occurrence of these tumors, nor was any one intercostal nerve significantly involved more frequently than any other. The symptoms tend to be fairly characteristic. The patient

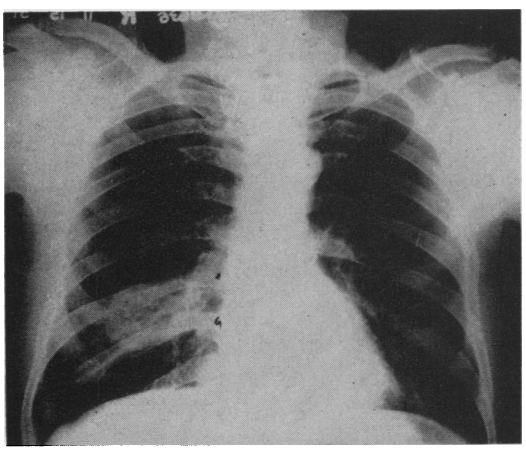

FIG. 5. Roentgenogram of chest revealing a mass in the chest opposite the eighth and ninth ribs posteriorly.

usually complains of pains in the chest wall, at the site of the tumor, and radiation of pain along the course of distribution of the involved nerve. The pain is frequently severe and persistent, and may be present for a long time before the patient seeks medical aid. There may be local tenderness to pressure at the site of the tumor. In some cases a portion of the tumor may be palpable through the intercostal space.

In cases where the tumor assumes massive size, symptoms may be produced as a

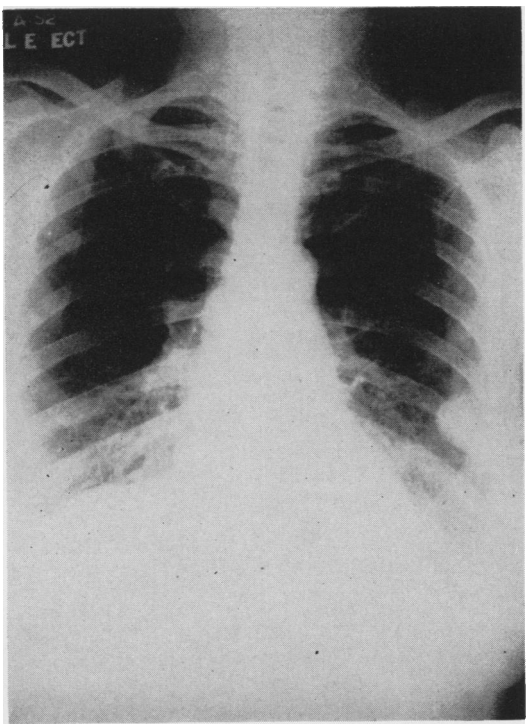

FIG. 6. Roentgenogram of the chest showing a circumscribed neoplasm in the region of the left eighth posterolateral rib. Same case as Figure 7.

result of compression and/or impairment of function of neighboring tissues. The diagnosis can and has been made preoperatively.[1] The roentgenograms of the chest are fairly characteristic of this condition. A circumscribed dense area is found within the chest wall, as shown in Figures 3, 5, 6, 8 and 9. Institution of a partial pneumothorax will help delineate the tumor and help localize the site of origin of the tumor. Tomographic studies of the chest wall may help localize the site of the tumor. At times the growth is round, oval, and even irregular in shape, and may erode the adjacent rib (Fig. 4).

TABLE I. *Collected Cases of Benign Neurogenic Tumors of the Chest Wall.*

No.	Source	Age Sex	Diagnosis	Location	Symptoms	Remarks
1.	Palugyay 1920[23]	59 F	Neurofibroma	Intercostal nerve, left chest wall	Pain	Surgical excision. Operative death.
2.	Harrington 1929[11]	35 F	Neurofibroma	Seventh to tenth ribs, posterior auxillary line	Pain; tumor, palpable 8 years	Surgical excision. Recovery.
3.	Canigiani 1931[4]	38 F	Neurofibroma	Sixth intercostal space, left lateral chest wall	Pain	Surgical excision, recovery. Rib erosion present. Small mass palpable in interspace. Von Recklinghausen's disease present.
4.	Kinebock 1932[20]	33 M	Neurofibroma	Seventh rib, left lateral chest wall, "apple size"	Pain	Surgical excision, recovery. Von Recklinghausen's disease present.
5.	Cutler 1936[8]	27 F	Neurofibroma	Eighth intercostal nerve, right posterior chest wall, 9×7.5×7.5 cm.	Pain 5 years	Surgical excision. Recovery.
6.	Ivanissevich 1938[17]	17 M	Neurilemoma (Schwannoma)	Ninth rib, posterolateral chest wall, 7×5×5 cm.	Pain 7 mos.	Surgical excision, recovery. Small mass palpable in interspace.
7.	Odessky 1941[22]	37 F	Neurilemoma (Neurinoma)	Left lateral upper chest wall	....	Surgical excision, recovery.
8.	D'Abreau 1947[1]	56 M	Neurofibroma	Eighth intercostal nerve in the right axillary line, 5×2.5 cm.	Pain	Surgical excision, recovery Preoperative diagnosis correct.
9.	D'Abreau 1947[1]	29 F	Neurofibroma	Third intercostal nerve, 4×3×2 cm.	Pain	Surgical excision, recovery.
10.	Tourcff 1949[27]	39 M	Neurofibroma	Fourth rib, right mid-axillary line, 7×4×3 cm.	Mass on x-ray; Pain	Surgical excision, recovery. Pain developed after tumor was detected in a routine x-ray of chest.
11.	Sommer 1950[25]	30 F	Neurofibroma	Fifth rib, right lateral chest wall, 5×5×2.5 cm.	Pain, 4 yrs.	Surgical excision, recovery.
12.	Blades 1950[4]	—	Neurofibroma	Chest wall	——	Surgical excision.
13.		—	Neurofibroma	Chest wall	——	Surgical excision.
14.		—	Neurofibroma	Chest wall	——	Surgical excision.
15.		—	Neurofibroma	Chest wall	——	Surgical excision.
16.		—	Neurilemoma (Perineural fibroblastoma)	Chest wall	——	Surgical excision.
17.	Ackerman 1951[2]	47 F	Neurilemoma ("ancient")	Left anterior interspace lateral to sternum	Palpable mass, 3 mos.	Surgical excision, recovery. Palpable mass in interspace.
18.	Ackerman 1951[2]	27 M	Neurofibroma	Left posterior chest wall, 8 cm.	None	Surgical excision, recovery. Cutaneous pigmentation.
19.	Authors' Case 1	42 F	Neurilemoma	Seventh rib, right lateral chest wall, 10×6.5×4 cm.	Pain 29 yrs.	Surgical excision, recovery. Rib eroded.
20.	Case 2	49 M	Neurofibroma	Eighth intercostal space, right lateral chest wall, 3×4×7 cm.	Pain 6 mos.	Surgical excision, recovery.
21.	Case 3	54 F	Neurofibroma	Eighth intercostal space, left lateral chest wall.	Pain 2 yrs.	Surgical excision, recovery.
22.	Case 4	67 M	Neurofibroma	Seventh intercostal space, right, 4 cm.	Pain 6 yrs.	Surgical excision, recovery.

CASE REPORTS

Case 1. H. H., a 42-year-old negress, entered the hospital in January, 1948, giving a 29-year history of pain in the right chest in the axillary area. The pain was intermittent, but tended to become more severe as time passed. In 1941, when she first consulted a physician for the pain in the chest, she was told that a roentgenographic examination of the chest revealed a tumor. Surgery was advised at that time, but the patient refused operation. Several months prior to entering the hospital in January, 1948, the pain became so severe that she was unable to work. At the time of admission to the hospital the physical examination revealed slight diminution of expansion of the right side of

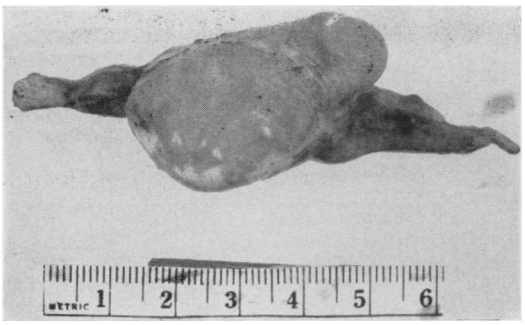

FIG. 7. Tumor removed from patient whose roentgenogram is seen in Figure 6. Note part of the intercostal nerve entering and leaving the body of the neoplasm.

course was uneventful. When seen in the clinic two years after the operation, there were no signs of recurrence.

Case 2. J. M., a 49-year-old man was admitted to the hospital in January, 1952, complaining of vague pains in the chest, but most commonly present on the anterior aspect of the right side. The onset of pain was approximately 6 months before entering the hospital. The physical examination of the chest was essentially negative. Roentgenograms of the chest revealed a density in the right lung field adjacent to the eighth and ninth ribs posteriorly (Fig. 5). After introducing some air into the pleural cavity it was noted that the neoplasm arose from the chest wall. The nature of the tumor

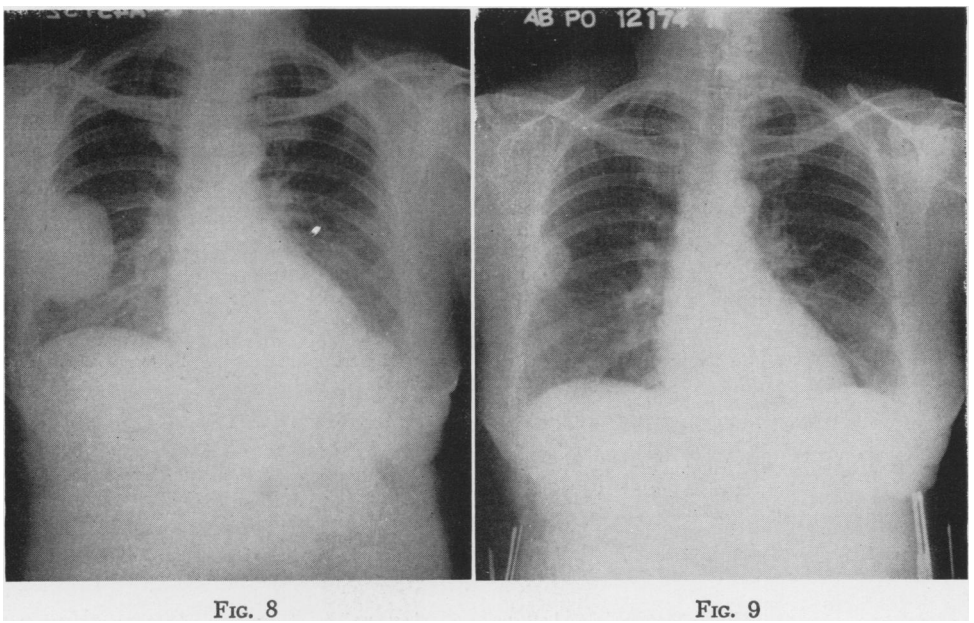

FIG. 8 FIG. 9

FIG. 8. A circumscribed mass is present in the seventh intercostal space on the right side.
FIG. 9. Same case as noted in Figure 8, but four years later.

the chest. A roentgenographic examination (Fig. 3) of the chest revealed a large circumscribed tumor in the region of the seventh to the ninth posterior ribs on the right side. Surgical removal of the tumor was instituted several days after admission to the hospital. A well encapsulated tumor (Fig. 4) measuring 10 x 6.5 x 4 cm., and the partially eroded seventh rib, were removed. The tumor was firm, and was red, orange and yellow in color on its surface and on cross-section. The outer surface was redder than the cut surface. At one end of the tumor a nerve could be seen entering the tumor. The histopathologic diagnosis was neurilemoma (Fig. 2). The patient's postoperative was uncertain. Surgical removal of the tumor was advised and instituted soon after entering the hospital. At operation a tumor was found in the region of the eighth intercostal nerve and outside of the parietal pleura. However, it was so firmly attached to the pleura that removal of the tumor could not be effected without removal of the locally adherent parietal pleura. The tumor was well encapsulated, pale yellow with areas of necrosis and recent hemorrhage visible on its cut surface. The microscopy showed the tumor to be a neurofibroma (Fig. 1). The patient had an uneventful postoperative course without recurrence of the tumor when last seen seven months after the operation.

Case 3. S. C., a 54-year-old woman, was admitted to the hospital in February, 1952, because of persistent pain in the left side of the chest for about 2 years. The pain was intermittent but progressive, and always in the same area. In recent months the patient developed a cough with expectoration of mucoid material. A roentgenographic examination of the chest revealed a mass in the region of the eighth rib in the posterior axillary area of the left side of the chest (Fig. 6). Surgery was advised and instituted soon thereafter. At operation, a tumor mass was found arising from the eighth intercostal nerve (Fig. 8), with the nerve seen going into and leaving the tumor mass. The tumor and intercostal nerve in proximity to the neoplasm were removed. The growth measured 4 x 2.5 x 2 cm. and was yellowish-pink on its exterior. On cross section of the tumor it was found to be of a like color and granular in appearance. The patient's postoperative course was uneventful. The histopathologic diagnosis was "neurofibroma."

Case 4. G. H., a 67-year-old man, was first seen at the hospital in November, 1946, complaining of pains on the right side of the chest of about 2 years' duration. The pain was more annoying than severe. Roentgenographic studies of the chest at this time disclosed a small circumscribed density in the region of the seventh intercostal space, laterally, on the right side of the chest (Fig. 8). Surgery was advised but the patient refused. He was seen thereafter from time to time; operation was advised and refused each time the patient was seen. In September, 1950, the pain became quite severe and the patient requested surgery. Roentgenographic studies of the chest showed the tumor to be about twice the size noted in the 1946 roentgenograms (Fig. 9). At operation a mass was found in the seventh intercostal nerve which measured 5 x 4 x 2.5 cm. The tumor was excised with part of the intercostal nerve; the postoperative course was uneventful. The histopathologic diagnosis was "neurofibroma."

DISCUSSION

Benign tumors of the intercostal nerves present a fairly characteristic symptom complex—local pain, and a fairly uniform roentgenographic pattern—a localized opacity within the chest wall. On rare occasions the corresponding rib may show pressure necrosis or erosion. The therapy unquestionably is surgical excision. The procedure is not formidable and is indicated not only to relieve the symptoms, but to remove a tumor which is potentially malignant or one that is already malignant, for a distinct differentiation cannot be made until representative tissue is examined under the microscope.

Resumé: Eighteen cases of benign neurogenic chest wall tumors which were subjected to surgery were collected from the literature. To these we added four cases not previously reported. A typical clinical picture is presented which should enable one to establish the correct diagnosis preoperatively. Surgical removal is advised in all similar cases.

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